Spontaneous Splenic Rupture in Mononucleosis

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• **Infectious mononucleosis is a disease entity with many and various manifestations. Suspicion of the disease is of the utmost importance. When indicated, adequate tests often repeated must be made to prove or disprove a suspicion of infectious mononucleosis. The disease is usually a benign condition; one of the more serious complications is "spontaneous" splenic rupture. Whether rupture is indeed spontaneous or is caused by relatively light trauma to a weakened organ is debatable. Splenectomy is the treatment for this complication; it does not cure the underlying disease. Once the diagnosis of infectious mononucleosis has been established, abdominal examinations should be limited and cautious.**

INFECTIOUS MONONUCLEOSIS has been described as a protean disease which involves the reticuloendothelial system. It may affect the entire body with equal intensity or be manifest in symptoms relating to one or a few structures of this system. The onset is usually associated with malaise, upper respiratory tract infection, weakness, low grade fever, mild pharyngitis and enlargement of lymph nodes. Chemotherapy helps little and there is a lingering illness of longer than is normally expected. Approximately seven to ten days after onset, the heterophil antibody reaction becomes positive in significant diagnostic dilutions. If the disease is suspected yet not immediately proved by laboratory aids, tests for it should be repeated at regular intervals. It is important to suspect the disease when the lymph nodes are enlarged, for correct diagnosis greatly decreases the mortality. In the majority of cases, if properly treated with rest and symptomatic medication, it is an innocuous and benign condition. Once therapy is begun, the patient has symptomatic improvement and is likely to insist on resuming normal activities. The physician must not be influenced by the wishes of the patient. In certain instances the disease may be fulminating and fatal.

Spontaneous splenic rupture is one of the rarer complications. It occurred in two patients who were treated at Norton Air Force Base Hospital, San Bernardino, California. One of the patients was observed by the authors (Case 2). The report of Case 1 was abstracted from hospital records.

CASE 1. A 31-year-old male officer was admitted to the hospital on April 2, 1954. Ten days before admission the patient noticed the onset of nocturnal chills, fever and myalgia; and two days before admission, generalized lymph node enlargement and pronounced malaise developed.

Upon examination the patient was observed to be well developed and well nourished, in no acute distress, well oriented and cooperative. The blood pressure was 138/74 mm. of mercury, the pulse rate 88, the temperature 98.6°F. Diffuse pharyngitis, pronounced enlargement of cervical lymph nodes and moderate axillary lymph node enlargement were noted. No abnormalities were noted in the heart and lungs. The liver, palpable one and a half fingerbreadths below the right costal margin, was firm and slightly tender. The splenic edge was barely palpable.

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Results of urinalysis were within normal limits. Leukocytes numbered 18,200 per cu. mm. of blood —17 per cent neutrophils, 30 per cent lymphocytes, and 53 per cent abnormal lymphocytes. Erythrocytes numbered 4.2 million per cu. mm. and the hematocrit was 36 per cent. The sedimentation rate was 26 mm. in one hour. Heterophil reaction was positive in 1 to 1,792 dilution.

The patient was kept in bed and was given symptomatic treatment. He became quite nauseated on the morning of April 4 and had several bouts of vomiting. Several hours later the patient began to complain of intermittent pains in the left upper quadrant of the abdomen, which became progressively worse during the night.

On April 5 the abdomen was moderately distended with shifting areas of dullness to percussion in the flanks and positive fluid waves. Leukocytes numbered 16,050 per cu. mm.—29 per cent neutrophils and 40 per cent lymphocytes (30 per cent reported as abnormal). Erythrocytes numbered 2.6 million per cu. mm. and hemoglobin content was 9.5 gm. per 100 cc. The hematocrit was 26 per cent. A diagnosis of "acute surgical abdomen" was made. The abdomen was opened and the spleen was found to be enlarged and ruptured. It had become partially walled off by the stomach, colon and omentum. The spleen was removed.

The pathological report stated that the gross specimen consisted of a spleen 15 x 10 x 6 cm., weighing 432 gm. The capsule was almost completely fallen away, leaving the underlying pulp naked. On section, the specimen was found to be extremely pulpy with a loss of the usual architectural features.

Upon microscopic examination the sinusoids were observed to be filled with cells. Normal and atypical lymphocytes were seen. There were numerous areas where the spleen showed microscopic cleavage

planes and these areas were also filled with a similar cell in a protean coagulation. The pathological diagnosis was infectious mononucleosis of the spleen with rupture.

The patient made an uneventful postoperative recovery and was discharged on April 26, 1954. He was readmitted on June 7, 1954, with complaint of increasing fatigue, bloating and anorexia. Seven days previously a low grade fever had developed. At the time of admission the sclerae were decidedly jaundiced. A diagnosis of homologous serum hepatitis secondary to blood transfusion was made. Conservative treatment was given and the patient recovered. Thereafter he remained asymptomatic. The interval between the blood transfusion and the onset of symptoms on the second admission was 50 days—a relatively short incubation period for homologous serum hepatitis. The second illness may have been a relapse of infectious mononucleosis. Clinical hepatitis with jaundice is not an infrequent complication of infectious mononucleosis, as the liver is richly endowed with Kupffer cells, a part of the reticuloendothelial system.[3]

CASE 2. A 23-year-old white male airman was admitted to the hospital on December 11, 1954. A month previously the patient had had a mild upper respiratory tract infection that lasted four or five days. Three weeks later, a week before admittance, while engaged in a moderately strenuous training program, he had fallen several times, striking his abdomen against a pack that he carried. Four days before admission a nonproductive cough associated with weakness and extreme malaise developed. While taking a shower an hour before hospitalization, the patient had sudden, severe pain in the left upper quadrant of the abdomen. The pain was accentuated by coughing and deep inspiration.

Upon examination the patient was observed to be well developed and well nourished, well oriented and cooperative. The blood pressure was 126/70 mm. of mercury, the pulse rate 104, respirations 20 per minute and the temperature 100.2°F. The skin was pale, with cold, clammy perspiration. The posterior pharyngeal wall was of a dull red hue. No enlargement of lymph nodes was noted in the cervical or axillary regions. The heart and lungs were normal. Moderate voluntary splinting of the muscles of the upper abdomen was noted. No masses were palpated. The outline of the spleen could not be felt.

Leukocytes numbered 10,830 per cu. mm.—neutrophils 40 per cent, lymphocytes 46 per cent, abnormal lymphocytes 13 per cent, basophils 1 per cent. Erythrocytes numbered 4.3 million per cu. mm. and the hemoglobin content was 13.8 gm. per 100 cc. Results of urinalysis were within normal limits. The heterophil titer was positive in 1 to 3,584 dilution. Guinea pig absorption tests were specific for infectious mononucleosis during this time.

A diagnosis of infectious mononucleosis with possible "spontaneous" splenic rupture was considered. No abnormalities were noted in x-ray films of the abdomen and chest taken at the time of admission.

Six hours after admission, the patient complained of increasingly severe pain in the left upper quadrant of the abdomen. The patient was observed closely and repeatedly examined. The blood pressure and pulse remained stable. Left upper abdominal pain continued but it was intermittent and decreasing in severity. Blood examinations were done repeatedly and the hematocrit and hemoglobin content slowly decreased. In a plain film of the abdomen taken 24 hours after admission, mild gastric dilatation and the presence of a mass displacing the stomach in the left upper quadrant were observed. The patient was given a transfusion of 500 cc. of whole blood and pronounced symptomatic improvement occurred immediately.

On December 14, 1954 the patient complained of a sudden severe pain in the left upper quadrant of the abdomen, associated with pain in both shoulders. He was nauseated and became cold and clammy. The blood pressure was 90/60 mm. of mercury and the pulse rate 130. Transfusion of whole blood was started and the patient was taken to surgery. At operation a large walled-off mass was found in the left upper quadrant of the abdomen. In the left subdiaphragmatic space there were multiple old clots and a pulpy splenic mass which was bleeding from multiple rents. The splenic capsule was almost completely avulsed.

The pathological report said that the specimen consisted of a spleen weighing 310 gm. and measuring 15 x 13 x 5.5 cm. The external surface was pale brown. The gastric surface of the spleen was interrupted by numerous large, irregular gaping rents extending the entire length of the spleen. There were numerous dark red blood clots that measured up to 10 cm. in greatest dimension. Upon microscopic examination it was observed that the usual architectural pattern was greatly distorted by diffuse hyperplasia of the reticuloendothelial elements. The usual germinal centers were reduced in number and size. Within the sinusoids there was a typical cell that is seen in infectious mononucleosis—a large oval cell with pale cytoplasm and oval vesicular nuclei in which the chromatin is rather prominently clumped. Also within the sinusoids there were numerous polymorphonuclear cells, eosinophils, macrophages and a number of metamyelocytes. The diagnosis was infectious mononucleosis with splenic involvement and recent rupture of the spleen.

Postoperative recovery was uneventful. After ten days he was placed at complete bed rest for a period of one month as treatment of infectious mononucleosis. Two weeks following the operation heterophil reaction was positive in 1 to 890 dilution. Six weeks later positive reaction occurred at 1 to 224 dilution. Results of liver function tests were within normal limits. The patient was then completely asymptomatic. Heterophil reaction and liver function tests were done at monthly intervals for a period of six months with the thought that if nausea,

vomiting, anorexia or malaise developed again and were associated with a rise in the heterophil titer, the patient would be returned to bed rest.

COMMENT

The mortality rate associated with spontaneous splenic rupture in infectious mononucleosis is extremely high. In 23 cases reported, there have been five deaths or a mortality rate of 22 per cent. Even more impressive was the 40 per cent mortality rate in cases in which infectious mononucleosis was not diagnosed preoperatively or before death.[5] A preoperative diagnosis of infectious mononucleosis is of prime importance in decreasing the mortality rate. Since 1932, when Paul and Bunnell[4] discovered the presence of agglutinins and hemolysins for sheep erythrocytes in the serum of patients with infectious mononucleosis, the diagnosis of this disease has been on a firmer foundation.

Once the diagnosis has been established, the ever present possibility of splenic rupture must be borne in mind. The question as to whether the spleen actually ruptures spontaneously has been raised many times. There is extensive loss of normal splenic architecture in infectious mononucleosis. In the capsule and trabeculae considerable cellular infiltration occurs, with edema and so-called cloudy swelling. The basic structure of the spleen is greatly weakened. There is no doubt that the diseased organ is much less likely to withstand trauma than is a normal spleen.[5]

In Case 2 there was definite history of repeated concussive trauma. In Case 1 the diagnosis of infectious mononucleosis was made immediately, but even so it is probable the abdomen was palpated many times in the course of examination. In cases in which definite diagnosis is not made promptly, the patient undergoes many abdominal examinations for diagnosis. Frequently in reported cases, the onset of severe abdominal pain was related to trauma and stress such as that associated with vomiting and straining during bowel movement.[2] It would seem possible that even minor trauma of this kind might have relatively great effect on a severely diseased spleen. Once the diagnosis of infectious mononucleosis has been made, examination of the abdomen should be minimal, cautious and gentle.

Smith and Custer[5] noted that rupture of the spleen takes place at least three weeks after onset of the infectious mononucleosis. Presumably it is during this period that the capsular and trabecular changes progress to the point of rupture either "spontaneously" or through slight trauma. The symptoms of onset of "spontaneous" rupture of the spleen in infectious mononucleosis are quite variable. Rupture may be associated with onset of severe pain in the left upper quadrant of the abdomen, which may or may not be constant and accompanied by signs of intense shock. Gastric dilatation and displacement of the stomach by a soft tissue mass from the left upper quadrant as seen on a plain film are highly suggestive evidence. It is important to repeat the hemoglobin and hematocrit determinations for signs of slow bleeding which may not be clinically evident. It has been noted by various observers that splenic rupture was followed by reversal of the cell differential usual in infectious mononucleosis, with leukocytes and neutrophils rather than lymphocytes becoming predominant.[6] This was not noted in the two cases herein reported. A further aid in diagnosis is paracentesis. If blood is obtained, the diagnosis is supported; but even if it is not, the suspicion of ruptured spleen cannot be allayed, for the organ may be walled off. Once the diagnosis is made, operation should be done immediately.[1]

Splenic rupture with hemorrhage progresses in two ways. One is by immediate hemorrhage with exsanguination and death. This may occur within three hours after the onset of acute symptoms of abdominal pain. The other way is by delayed hemorrhage—the delay being caused by tamponade, as happened in both the cases reported herein. In each, at the time of operation, it was noted that a partial walling-off had taken place. Tamponade occurs in the following manners: Simple rupture with clot formation, simple or multiple ruptures with omental plug, simple or multiple ruptures with walling-off by the stomach, colon and omentum.[7]

It is impossible to estimate the incidence of infectious mononucleosis and the complication of splenic rupture. It is probable that it is rather rare.[3] In the differential diagnosis of acute abdominal pain it must always be considered. This condition may simulate such entities as a ruptured peptic ulcer, acute cholecystitis, acute hemorrhagic pancreatitis, acute appendicitis with peritonitis, acute mesenteric thrombosis and acute intestinal obstruction.

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